CASE REPORT
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ENDOSCOPIC TREATMENT OF THORACIC SCHWANNOMATOSIS: A CASE REPORT

TRATAMENTO ENDOSCÓPICO DE SCHWANNOMATOSE TORÁCICA: RELATO DE CASO

TRATAMIENTO ENDOSCÓPICO DE SCHWANNOMATOSIS TORÁCICA: REPORTE DE CASO

Ramon Venzon Ferreira[1], Diogo Martins Maldonado[1], Kelsen de Oliveira Teixeira[1,2,3], Cesar Luiz Bertonha[1], Esthael Cristina Querido Avelar Bergamaschi[1,2,3], Leonardo Ribeiro Nascimento[1,2,4], Sonja Ellen Lobo[2], João Paulo Machado Bergamaschi[1,2]

1. Universidade de São Paulo (USP-RP), School of Medicine, Specialization Course in Spinal Endoscopy Surgery, Ribeirão Preto, SP, Brazil.
2. Instituto Atualli, São Paulo, SP, Brazil.
3. Atualli Spine Care Clinic, São Paulo, SP, Brazil.
4. Atualli Spine Care de Franca, Franca, SP, Brazil.

ABSTRACT

Schwannomatosis, also known as neurofibromatosis type III, is a rare condition characterized by the development of multiple schwannomas along peripheral nerve sheaths. Although open surgery remains the standard treatment, the evolution of minimally invasive techniques has expanded the indications of endoscopic spine surgery, including for benign expansile lesions. We present the case of a 76-year-old female with thoracic pain, lower limb neurological deficits, and imaging findings suggestive of thoracic schwannomas associated with a disc herniation. The patient underwent a posterolateral extraforaminal endoscopic resection of both the disc herniation and the adjacent nerve sheath tumor. The procedure was uneventful, with partial improvement in pain and motor function. Further investigation confirmed the diagnosis of Schwannomatosis with multiple lesions throughout the spinal axis. Endoscopic surgery proved to be a feasible and safe approach for decompression in cases of isolated thoracic schwannomas. Further studies are warranted to confirm its role in the treatment of spinal neoplastic conditions. ***Level of Evidence IV; Case Report.***

Keywords: Neurilemmoma; Neurofibromatosis Type 3; Constriction, Pathologic; Minimally Invasive Surgical Procedures; Spine.

RESUMO

A Schwannomatose (SCH), ou neurofibromatose tipo III, é uma condição rara caracterizada pelo desenvolvimento de múltiplos schwannomas na bainha nervosa periférica. Embora a cirurgia aberta seja a principal abordagem descrita na literatura, os avanços nas técnicas minimamente invasivas têm ampliado as indicações da cirurgia endoscópica da coluna vertebral, inclusive para lesões expansivas benignas. Apresentamos o caso de uma paciente de 76 anos com quadro de dor torácica, déficits neurológicos em membros inferiores e lesões sugestivas de schwannomas torácicos associadas a hérnia discal. A paciente foi submetida a ressecção endoscópica póstero-lateral extraforaminal da hérnia discal e da lesão expansiva. O procedimento transcorreu sem intercorrências, com melhora parcial da dor e da função motora. Investigações complementares confirmaram o diagnóstico de SCH, com múltiplas lesões ao longo do eixo vertebral. A cirurgia endoscópica demonstrou ser uma abordagem viável e segura para descompressão em casos de schwannomas torácicos isolados. Estudos adicionais são necessários para consolidar sua aplicação no manejo de doenças neoplásicas da coluna. ***Nível de Evidência IV; Relato de Caso.***

Descritores: *Neurilemoma; Neurofibromatose Tipo 3; Constrição Patológica; Procedimentos Cirúrgicos Minimamente Invasivos; Coluna Vertebral.*

RESUMEN

La Schwannomatosis, también conocida como neurofibromatosis tipo III, es una enfermedad rara caracterizada por el desarrollo de múltiples schwannomas en las vainas nerviosas periféricas. Aunque la cirugía abierta sigue siendo el tratamiento estándar, los avances en técnicas mínimamente invasivas han ampliado las indicaciones de la cirugía endoscópica de columna, incluso para lesiones expansivas benignas. Se presenta el caso de una paciente de 76 años con dolor torácico, déficits neurológicos en miembros inferiores y lesiones compatibles con schwannomas torácicos asociados a hernia discal. La paciente fue sometida a resección endoscópica extraforaminal posterolateral del fragmento discal y de la lesión expansiva. La cirugía transcurrió sin complicaciones, con mejoría parcial del dolor y de la función motora. La investigación complementaria confirmó el diagnóstico de Schwannomatosis, con múltiples lesiones en la columna vertebral. La cirugía endoscópica demostró ser una alternativa viable y segura para la descompresión en casos de schwannomas torácicos aislados. Se necesitan estudios adicionales para consolidar su aplicación en el tratamiento de afecciones neoplásicas de la columna. ***Nivel de Evidencia IV; Relato de Caso.***

Descriptores: *Neurilemoma; Neurofibromatosis Tipo 3; Constricción Patológica; Procedimientos Quirúrgicos Mínimamente Invasivos; Columna Vertebral.*

Study conducted by the Clínica Atualli Spine Care. Address: Rua Bela Cintra, 539, 2nd floor, Consolação, São Paulo/SP, Brazil.
Correspondence: João Paulo Machado Bergamaschi. Instituto Atualli, 539, Rua Bela Cintra, 4º andar, Consolação, São Paulo, SP, Brazil. jberga@clinicaatualli.com.br

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INTRODUCTION

Spinal endoscopic surgery has undergone a remarkable expansion in recent years, consolidating itself as a minimally invasive technique of high precision, with well-established applications in the treatment of disc hernias and lumbar and cervical stenosis. With the advancement of anatomical knowledge, the development of specific instruments and the accumulation of experience by surgeons, new indications have been explored, including in regions traditionally considered challenging, such as the chest spine. Although the literature on these approaches remains limited, a growing number of reports highlight their potential to achieve effective neural decompression with minimal morbidity[1,2].

Historically, the surgical treatment of intradural expansive lesions, such as nerve vagina tumors, was restricted to open and aggressive techniques, requiring extensive approaches and, often, prolonged hospitalizations. The possibility of treating these lesions endoscopically was considered, until recently, unattainable. However, the evolution of extraforaminal post-lateral approaches, coupled with direct visualization and adequate hemostatic control, has allowed effective interventions even in more complex cases. These advances pave the way for a new frontier of spinal endoscopy, where tumors previously considered inaccessible by endoscopy are now safely addressed[3,4].

The aim of this article is to report a clinical case of schwannomatosis (SCH) with thoracic impairment, undergoing surgical treatment through endoscopic approach, highlighting the technical aspects of surgery, the clinical-radiological findings, the functional outcome and the diagnostic considerations related to the rare etiology of the disease.

REPORT CASE

A 76-year-old female patient with no known comorbidities, no history of trauma, and no clinical or laboratory evidence of infection presented with moderate-intensity thoracic back pain associated with progressive bilateral lower limb weakness, more pronounced on the left side. Over a period of approximately six months, she developed progressive difficulty with ambulation and eventually became dependent on crutches for mobility.

The pain was quantified using the Visual Analog Scale (VAS), with a score of 7/10 in the thoracolumbar region. The functional disability was evaluated by the *Oswestry Disability Index* (ODI), resulting in 60%, characterizing intense functional limitation. On physical examination, the patient exhibited diffuse hypoesthesia in the lower limbs, an unsteady gait, bilateral muscle weakness predominantly affecting the left lower limb, and diminished bilateral patellar and Achilles reflexes.

Magnetic resonance imaging (MRI) of the chest spine revealed multiple expansive lesions with characteristics of nerve membrane formations, isointense in T1 and hyperintensive in T2, located in T10–T11 and adjacent segments, suggesting multiple schwannomas. These formations occupied intraspinal space and were insinuated by the neural forum, exerting significant compression on the neural structures. Concomitantly, posterolateral disc extrusion was observed to the left at the level of T10–T11, also contributing to the narrowing of the canal and radical compression. However, the expansive pattern of tumor lesions was the main finding responsible for the deformation of the tissue bag and the clinical picture of progressive neurological impairment (Figures 1 and 2).

In view of the clinical picture and image findings, an endoscopic post-lateral extraforaminal approach was chosen on the left, aiming at the resection of the cystic lesion associated with the removal of the herniated fragment from the adjacent intervertebral disc, which was insinuated by the ipsilateral foramen. The cutaneous incision was carried out at 8 cm from the middle line, with about 7 mm of extension. The procedure was conducted under local anesthesia (lidocaine 1% without vasoconstrictor) associated with conscious sedation.

The initial puncture was guided by fluoroscopy, targeting the lateral part of the joint facet of T10–T11 on the left. After confirmation

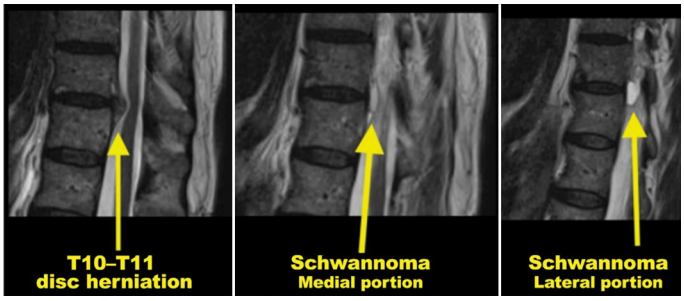

Figure 1. Images in sagittal cuts of the magnetic resonance of the chest spine, evidencing multiple expansive formations hypointensive in T1 and hyperintensive in T2, located in the topography of the nerve pelvis, compatible with schwannomas. Concomitant disc hernia is also observed at level T10–T11.

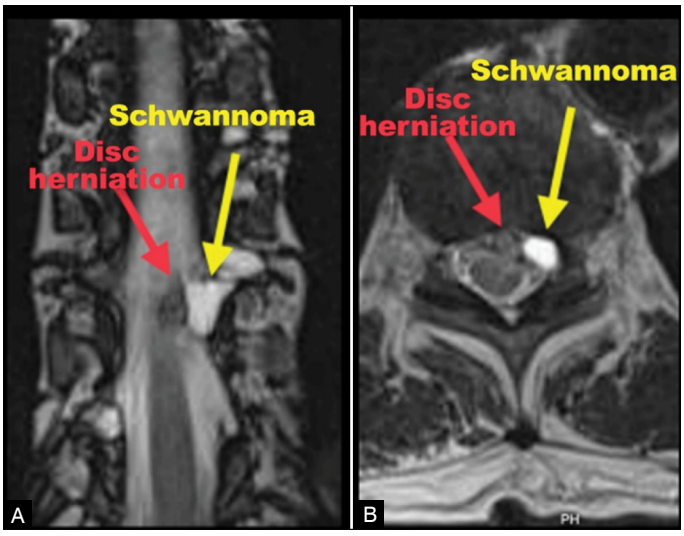

Figure 2. Images of magnetic resonance imaging in coronal (A) and axial (B) cuts, demonstrating vertebral canal stenosis due to expansive nerve pelvis injury associated with discal hernia, causing spinal cord and nerve root compression at T10–T11 level.

of the positioning of the punching needle and guide wire, sequential dilatators followed by the working canula were introduced. The endoscope used presented a work channel of 4.3 mm and a lens with an angle of 30 degrees, providing adequate visualization of the surgical field.

After initial hemostasis with bipolar forceps and soft tissue resection, foraminoplasty was performed for exposure of disc hernia and cystic lesion. Endoscopic discectomy with decompression of the medullary canal, followed by complete resection of intracanal tumor lesion (Figures 3 and 4). The medulla was visualized with no signs of residual compression. After hemostatic control, the endoscopic system was removed and the skin wound sutured with a 3/0 single nailon point. The procedure lasted approximately 45 minutes, with no anesthetic or clinical interactions.

After three months of outpatient follow-up, significant improvement in dorsalgia was observed (VAS 3/10) and significant reduction in ODI (26%), indicating moderate functional disability. There was also an improvement in the pattern of walking, although the patient still showed flambant walking, with changes in pelvic control in the balance phase and residual motor deficit in the lower limbs.

In view of the persistence of neurological dysfunction, a review of the imaging examinations and anatomopathological analysis was carried out, which confirmed the diagnosis of perineural pelvis tumor (schwannoma) in multiple vertebral segments (Figure 5). Electroneuromyography (ENMG) revealed bilateral peripheral neurophysiological impairment, with reduced amplitudes and driving speeds, especially in dorsiflexors of the feet.

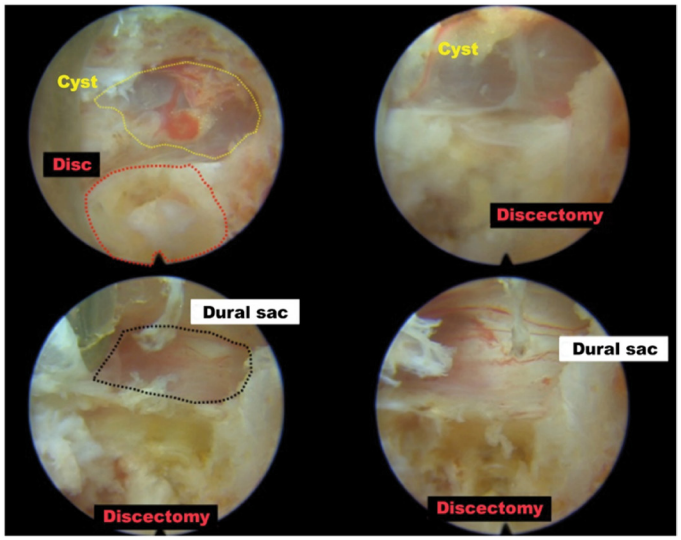

Figure 3. Intraoperative endoscopic images demonstrating exposure of the intervertebral disc and tumor cystic lesion, with direct decompression of the spinal cord after resection of the tumor mass and the herniated disc fragment.

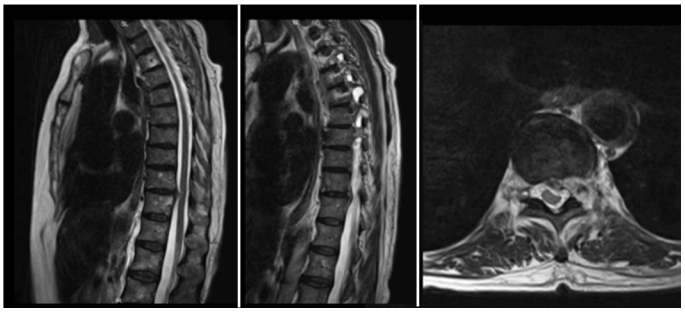

Figure 4. Postoperative magnetic resonance imaging in sagittal and axial cuts showing satisfactory decompression of the spinal cord at the operated level, after resection of schwannoma and discal hernia.

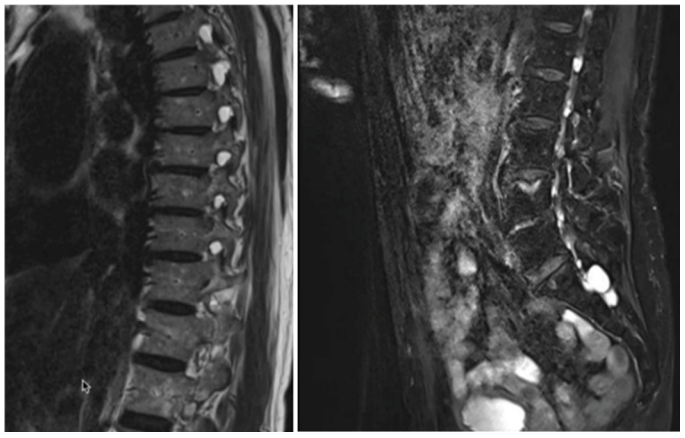

Figure 5. Magnetic resonance imaging of the chest and lumbar spine in sagittal cuts, evidencing multiple expansive lesions in different chest and lumbar segments, compatible with diagnosis of SCH (Neurofibromatosis type 3).

The clinical and complementary research was expanded, excluding differential diagnoses such as lumbosacral root compression, myelopathy, equine tail syndrome and degenerative diseases of the anterior horn, including amyotrophic lateral sclerosis. The pattern of walking observed in the patient was also a consequence of the severe manifestation of falling foot, resulting from peripheral neuropathy. The impairment of the afferent and eferent pathways, especially of the proprioceptive and distal motor sensitive fibers, resulted in loss of postural balance and significant changes in deambulation.

Based on the clinical, electrophysiological, radiological and anatomopathological findings, the diagnosis of SCH (neurofibromatosis type 3) was confirmed, a rare condition characterized by multiple nerve vagina tumors, neuropathic pain and progressive neurological deficits.

After the initial minimally invasive surgical approach, the patient was referred to follow-up with a team of neurology, motor physiotherapy and multidisciplinary pain treatment. Considering that the main compressive lesion had been completely removed and the remaining had extensive distribution (lumbar, dorsal and cervicotoracic regions), conservative behavior was chosen against the other tumor formations. The patient remains in clinical follow-up with adequate pain control (VAS 2), active participation in functional rehabilitation and specialized neurological follow-up, reinforcing the importance of an individualized approach in cases of schwannomatosis with multifocal spinal impairment.

DISCUSSION

Spinal canal stenosis may have a congenital or acquired etiology. Among the acquired forms, degenerative, traumatic, iatrogenic, and neoplastic causes are the most common[2]. In the context of neoplastic etiologies, nerve vagina tumors, in particular schwannomas, play a significant role by occupying space in the vertebral canal and generating neural compression. These tumors are often associated with SCH, also called neurofibromatosis type III, a rare condition belonging to the group of neurofibromatosis and affecting approximately five thousand Brazilians (1:40,000 born)[3]. Despite the scarcity of epidemiological data on predominance by sex or age group of higher incidence, it is known that sporadic schwannomas, not linked to genetic syndromes, tend to manifest after the fourth decade of life.

The diagnostic criteria for SCH were established by MacCollin et al. (2005)[5], and include both defined and presumptive criteria. The defined diagnosis is confirmed when two or more schwannomas are identified, with anatomopathological evidence, and the absence of a tumor in the vestibular nerve, evidenced by an imaging examination performed after the age of 18 years[6,7].

Presumptive diagnosis can be considered in three situations: 1) when there are two or more schwannomas detected by biopsy, with or without signs of vestibular dysfunction after the age of 30; 2) when two or more schwannomas are found in delimited anatomical distribution, without vestibular symptoms, at any age, or 3) when there is a compatible family history, associated with suggestive clinical and radiological findings. Only about 15% of cases have a family character, usually associated with deletions or mutations in chromosome 22, especially involving the tumor suppressor gene SMARCB1 (hSnf5/INI1)[8].

These criteria are fundamental to differentiate SCH from other neurofibromatosis, especially type 2, and guide the clinical and genetic management of patients.

The histopathological differential diagnosis between schwannomas and neurofibromas is essentially based on the tumor growth pattern[9]. Schwannomas are encapsulated neoplasms, composed almost exclusively of Schwann cells, and grow eccentrically to the nerve, compressing it peripherally. On the other hand, neurofibromas present diffuse intraneural growth, involving the nerve in a concentrated way and being formed by Schwann cells, axons, fibroblasts and perineural cells in rich extracellular matrix. In cases of SCH, schwannomas often exhibit peritumoral edema, exuberant myxoid transformation and intraneural growth pattern, in addition to strong positivity for protein S-100, characteristic finding of these cells[10].

Symptomatology usually appears after the age of 30 and half of patients have changes in the neurological examination. Pain in the lower limbs is a common manifestation, often of difficult control[11]. Studies estimate that up to two-thirds of patients with SCH develop neuropathic pain associated with tumor appearance, although there is no direct correlation between the volume of lesions and the intensity of pain[12]. Motor deficits may also be present[12,13] reinforcing the multifactorial character of neurological impairment in these patients.

In the present case, in the face of advanced age, clinical manifestations and the findings of MRI, endoscopic surgical treatment was initially chosen for decompression of the vertebral canal, with removal of discal hernia and excision of the predominant perineural lesion. The procedure resulted in adequate decompression and initial clinical improvement; however, the persistence of neurological changes, such as walking disorders, muscle weakness and hyperreflexia, motivated extended diagnostic research in the postoperative period.

The association between such neurological findings, intraoperative aspects, images of adjacent levels and the anatomopathological outcome led to the targeted investigation, culminating in the diagnosis of SCH, a rare condition, often underdiagnosed and associated with chronic neuropathic pain. Among its distinctive histological characteristics, the peritumoral edema of the adjacent nerve[3] stands out, consistent with the pattern observed in this case.

Although ENMG is not part of the formal diagnostic criteria of SCH[14], its use has been essential to rule out differential diagnoses associated with lower limb weakness. The neurophysiological study demonstrated reduction of the amplitudes and speeds of neural conduction bilaterally, most evident in the dorsiflexors, in addition to abolishing H reflexes and F waves, findings compatible with diffuse peripheral neuropathy. Specific protocols for investigation of lumbosacral root compression, myelopathy, equine tail syndrome and anterior cornea diseases, including amyotrophic lateral sclerosis, were applied and discarded. The electrophysiological findings strengthened the diagnosis of peripheral polyneuropathy, evidencing the complexity of the case and the need for a multidisciplinary approach.

CONCLUSION

The use of the extraforaminal post-lateral endoscopic technique for resection of lesions associated with SCH has been demonstrated to be viable, safe and effective, even in cases with spinal cord compression. Good intraoperative visualization, adequate hemostatic control and favored postoperative recovery reinforce the potential of this minimally invasive approach in the management of benign expansive lesions of the nerve pelvis in the chest spine, even when associated with symptomatic disc hernia.

Despite the good results obtained in this report, it is fundamental to highlight the scarcity of specific literature on the use of spinal endoscopy in intracranial tumors. The consolidation of this technique for such indications depends on the conduct of more robust clinical studies, with a larger sample size and long-term follow-up, in order to validate its effectiveness, safety and definitive role in the therapeutic arsenal of spinal tumor diseases.

CONFLICT OF INTEREST

All authors declare no potential conflict of interest related to this article.

CONTRIBUTIONS OF THE AUTHORS

Each author contributed individually and significantly to the development of this article. RVF, DMM, KOT, LRN: conceptualization, methodology, writing - original draft; CLB, ECQAB: validation, data curation; SEL, JPMB: formal analysis, writing - review and editing.

DATA AVAILABILITY DECLARATION

The contents underlying the research are available in the manuscript.

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